

Emergency Room Protocol

**A handy reference guide to provide everything you need for
emergency room visits as a patient with a Mast Cell Disorder.**

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The Mastocytosis Society, Inc. also maintains a message only phone line that patients and health care professionals may call in an emergency and leave a message. A Board of Directors member will reply or contact the necessary physician specialist on their behalf.

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WHAT TO SAY UPON ARRIVING IN THE EMERGENCY ROOM: BRIEF SCRIPTS

1. If your problem is a full blown mast cell attack/mast cell degranulation/fainting/anaphylaxis:

I am ___ years old with a known systemic mast cell disorder, and I am having anaphylaxis. (Say this even if you have not experienced anaphylaxis before as any mast cell degranulation attack can result in full blown anaphylaxis.)

2. If your problem is something else, (virus, fracture, auto accident, etc) and your mast cells are already flaring up:

I am ___ years old with _____ (i.e. an injury to my left knee). I also have a systemic mast cell disorder, and I am already showing signs of mast cell degranulation, which can quickly turn into anaphylaxis. I need to be treated immediately to prevent that from happening.

3. If you have cutaneous disease but have systemic symptoms from the mast cell mediators released from your skin lesions:

I am ___ years old and I have cutaneous mastocytosis with systemic Effects from the release of mediators in my skin lesions. I am at high risk for anaphylaxis and need to be treated immediately to prevent that.



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Medical Emergency Response Plan for Systemic Mastocytosis, Mast Cell Activation, and Anaphylaxis

If the patient presents with flushing, rash, hives, swelling, abdominal pain, nausea, vomiting, shortness of breath, wheezing or hypotension, respond with the following:

Administer

- Epinephrine 0.3 cc of 1/1000 and repeat 3x at 5-minute intervals if BP < 90 systolic (0.1 cc for children under 12)
- Benadryl (Generic: diphenhydramine) 25-50 mg (12.5-25 for children under 12) orally, intramuscular or intravenously every 2—4 hours or Atarax (Generic: hydroxyzine) 25mg (12.5 mg for children under 12) orally every 2—4 hours
- Solu-Medrol (Generic: methylprednisolone) 120 mg (40 mg for children under 12) IV/IM
- Oxygen by mask or nasal canula 100%
- Albuterol nebulization

Call 911 and take the patient to the closest Emergency Room.

Pre-medication for major and minor procedures and for radiology procedures with and without dyes:

- Prednisone 50 mg orally (20 mg for children under 12) 24 hours and 1—2 hours prior to surgery
- Benadryl (Generic: diphenhydramine) 25-50 mg orally (12.5 mg for children under 12) or Atarax (Generic: hydroxyzine) 25 mg orally, 1 hour prior to surgery
- Zantac (Generic: ranitidine) 150 mg orally (20 mg for children under 12) 1 hour prior to surgery
- Singulair (Generic: montelukast) 10 mg orally (5 mg for children under 12) 1 hour prior to Surgery

Drugs to be avoided:

- Aspirin and non-steroidal anti-inflammatory medications
- Morphine, codeine derivatives
- Vancomycin

Recommend: Tylenol

The Mastocytosis Society thanks Dr. Mariana Castells for this emergency protocol.

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QUICK REFERENCE GUIDE: MEDICATIONS TO USE AND AVOID IN PATIENTS WITH MAST CELL DISEASE IN EMERGENCY SITUATIONS

Please note: Some of the Drugs to Avoid may be given if absolutely necessary, if given with a prep to stabilize mast cells. Please refer to one of our mast cell experts for instructions.

Medication Type	<u>AVOID THESE DRUGS</u>	Drugs that are typically tolerated
General Drugs	<ul style="list-style-type: none">- Alcohol- Amphotericin B- anticholinergic drugs- Dextran- Dextromethorphan- Ethanol- Polymyxin B- Quinine- Vancomycin IV- α-adrenergic blockers- β-adrenergic blockers	
Pain Medications	<ul style="list-style-type: none">- Opioid narcotics (may be tolerated by some individuals)- Toradol- Non-steroidal anti-inflammatory drugs (unless the patient is already taking a drug from this class)	<ul style="list-style-type: none">- Fentanyl (may require adjunct treatment with Zofran)- Tramadol
Muscle Relaxants	<ul style="list-style-type: none">- Atracurium- Doxacurium- D-tubocurarine- Metocurine- Mivacurium- Succinylcholine	<ul style="list-style-type: none">- Pancuronium- vecuronium

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QUICK REFERENCE GUIDE

Local Anesthetics	<ul style="list-style-type: none"> - Benzocaine - Chloroprocaine - Procaine - Tetracaine 	<ul style="list-style-type: none"> - Bupivacaine - Lidocaine - Mepicacaine - Prilocaine - Levobupivacaine - Ropivacaine
Intraoperative Induction Meds		<ul style="list-style-type: none"> - Ketamine - Midazolam - Propofol
Inhaled Anesthetics		<ul style="list-style-type: none"> - Sevoflurane

Please note: the majority of the information in this quick reference guide was taken from the article by Gould and Park, and placed into this quick reference guide for ease of use by ER practitioners.

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LABORATORY TESTS TO RUN IN THE EMERGENCY ROOM DURING AN ACUTE MAST CELL DEGRANULATION EPISODE OR ANAPHYLAXIS

1. Serum Tryptase-upon arrival in the ER and three hours later.
2. 24 hour urines for:
 - n-methyl histamine
 - prostaglandin D2 (PGD2)
 - prostaglandin 11 beta F2A (BPG-F2A)
3. Complete chemistry panel
4. CBC with differential

You MUST have your allergist or primary care provider sign the bottom of this form stating that he or she will be responsible for the follow-up on the 24 hour urine collections. Otherwise, the ER physicians will be reluctant to order them since they cannot be sure of follow-up care. Remember to contact your physician for follow-up after discharge.

I agree to provide follow-up care for my patient, _____,
And will obtain the results of the 24 hour urine collections that were initiated in the emergency room setting, and will provide appropriate care based on the results.

Printed Name of Physician

Signature of Physician

Date

Contact Address

Phone Number

Fax Number



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MEDIC ALERT AND OTHER MEDICAL JEWELRY

When deciding what to put on your medical jewelry, the first word should always be:

Anaphylaxis!

If, and only if, you are on a Beta blocker, add the following:

On Beta blockers-give glucagon with Epinephrine.

Followed by **systemic mastocytosis** or **systemic mast cell disorder**.

Do not put mast cell activation disorder yet until it is more uniformly recognized.

Then add (if you have them): **Drug Allergies**.

You probably have more than one, and since there will not be room to list them all, they will be kept at the Medic Alert headquarters. This is an advantage of choosing a medical jewelry company that stores data. An alternative is one that provides a USB flashdrive with all the details of your medical history, like American Medical.

Then add(if you have them): **Food Allergies**.

Then add any critical other allergies such as **Allergy to Insect Stings**.

Next add other illnesses, such as **diabetes**, **unstable vasospastic angina**, **Hashimoto's thyroiditis**, etc.

Thank you to the members of the TMS Medical Advisory Board who contributed their thoughts to this page.
Valerie M. Slee, Chair



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ADDITIONAL WAYS TO ADVOCATE FOR A CHILD IN THE EMERGENCY ROOM SETTING

1. Offer the copy of The Special Edition of The Mastocytosis Chronicles for Healthcare Professionals, and refer to the page on Pediatric Mastocytosis.
2. Explain that mast cells in cutaneous lesions release mediators that result in systemic symptoms, sometimes severe, such as flushing, nausea, vomiting, diarrhea, headaches, itching, difficulty concentrating, dizziness, etc. (add your child's symptoms here.)
3. Explain clearly how your child manifests early signs of deterioration or anaphylaxis, especially if it is not a typical presentation:
 - Does your child develop hives or swelling around the face or mouth? eyes, or develop flushing or pallor?
 - Does your child get an itchy, red rash (other than hives)?
 - Does your child develop a cough, especially one that can be staccato in nature in younger children, or may mimic their asthma cough
 - Does your child develop frequent sneezing and/or a runny nose?
 - Does your child exhibit shortness of breath?
 - Does your child complain of chest pain even in the absence of shortness of breath?
 - Does your child complain of a mouth or tongue that feels funny? (Note: this may happen well before any visible oral swelling can be recognized or appreciated on exam.
 - Does your child exhibit hoarseness or a change in voice?
 - Does your child clear his or her voice repetitively?
 - Does your child complain of trouble swallowing, or appear to be drooling excessively?
 - Does your child exhibit sudden abdominal pain?
 - Does your child develop nausea? vomiting? In some children, this may be the only initial symptom.
 - Does your child feel anxious, or tell you that something awful is happening?

You know your child best, so be sure to educate the ER about how to recognize early anaphylaxis in your child.

4. Make sure that all medications and IV additives are alcohol free.

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1. *Anaphylaxis*. Mayo Foundation for Medical Education and Research, 1998-2010, February 2009.
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WHAT YOU SHOULD KNOW ABOUT ANESTHESIA – IT COULD SAVE YOUR LIFE

Nancy Gould and Regis (Gigi) Park

INTRODUCTION

Surgery is a stressful experience. For a patient with mast cell disease, that stress is compounded by the possibility of complications including anaphylaxis, cardiovascular collapse, increased bleeding and even death. Therefore, general anesthesia is considered a high-risk procedure in patients with mast cell disease. It is critical that all members of the patient's operating team take proper precautions before, during, and after surgery to protect against potentially life-threatening mast cell activation.

PLANNING FOR SURGERY SHOULD BEGIN AS SOON AS THE NEED FOR SURGERY ARISES

It is imperative that good communication is established between the patient, referring physician, surgeon, anesthesiologist, nurses, and all others involved in the patient's care before, during, and after surgery. The surgeon must be aware of the patient's mast cell disease and inform themselves of the measures necessary to keep the patient as free of symptoms as possible. Additionally, the patient should contact the anesthesiologist assigned to their care as soon as possible after surgery has been scheduled. Both regional and general anesthetics can cause life-threatening complications. An experienced anesthesiologist is aware of medications known to cause mast cell degranulation and medications that stabilize mast cells. If the patient is satisfied that their anesthesiologist fully understands the importance of planning around the mast cell disease, it will go a long way toward calming the patient, which in turn may reduce mast cell mediator release.

Symptoms should be as well-controlled as possible in the days prior to surgery. The patient should carefully avoid known triggers of mast cell activity and should take their medications as prescribed. Those medications include H1 and H2 histamine receptor blockers such as Allegra (H1) and Zantac (H2). A mast cell stabilizer such as disodium cromoglycate or ketotifen, and medications targeting other mast cell mediators should be taken regularly if they are part of the patient's normal drug regimen.

The patient's complete medication list should be reviewed by the surgical team prior to surgery and any necessary medication changes should be thoroughly discussed with the patient. For example, if the patient regularly takes a medication in the family known as beta blockers for blood pressure or heart rate abnormality, a change to another drug should be considered well before surgery is scheduled. Beta blockers are generally avoided in people with mast cell disease undergoing surgery because they interfere with an important natural control of mast cell activation. These drugs may also interfere with the use of epinephrine, which may be required if the patient has a major release of mast cell mediators resulting in low blood pressure during surgery. Other drugs that may interfere with control of blood pressure during surgery must be carefully reviewed by the patient's physician before the surgery.

It may be necessary to perform a "graded challenge" procedure in the hospital under the supervision of an allergist and an anesthesiologist for certain medications if there is no history of exposure to that medication. This procedure usually starts with scratching the skin with a small amount of medication followed by injection of increasing amounts with careful monitoring after each injection. Resuscitation equipment and drugs including epinephrine must be readily available during the procedure.

PRECAUTIONS TAKEN IN THE HOURS PRIOR TO SURGERY WILL HELP THE PATIENT GO INTO SURGERY IN THE BEST POSSIBLE CONDITION

For pre-operative control of anxiety and the reduction of mast cell activity, drugs in the valium family (diazepam, midazolam, lorazepam) are usually effective. Some procedures require the patient not to take anything by mouth

including medications after midnight of the night before the surgery. In this case, H1 and H2 blockers should be administered intravenously prior to the surgery. The use of corticosteroids, such as prednisone, has also been suggested although there is no evidence that the short-term use of steroids reduces the ability of mast cells to release the chemicals contained in their granules. However, corticosteroids may reduce the extent of other inflammatory reactions that result from mast cell activation.

A tube may be inserted into an artery and attached to a device allowing the anesthesiologist to vigilantly monitor blood pressure without having to periodically inflate a blood pressure cuff. In addition, a tube is inserted into a vein and securely taped in place, with intravenous (IV) fluids running to keep the patient well-hydrated in all surgeries involving general anesthesia or conscious sedation. This tube will also make it possible to immediately administer any emergency medications that may be needed.

In the operating room, the patient should not be allowed to become either too cold or over-heated. Warm blankets should be used, and all IV fluids should be warmed before they are given. In addition, there should be a minimum of noise and bustle around the patient prior to the administration of the anesthetic in order to reduce the possibility of anxiety-triggered mast cell mediator release.

EMERGENCY SURGERY

An emergency situation may arise in which a person with mast cell disease requires immediate surgery. Wearing a MedicAlert bracelet could be a life-saver if this happens. Inscriptions vary depending upon the patient's specific needs, but generally include the patient's diagnosis and drug sensitivities. Emergency response team members can access more detailed information 24 hours a day, 7 days a week from the MedicAlert organization by dialling the toll-free number on the bracelet. MedicAlert is a non-profit organization serving patients world-wide. There are nominal membership and annual fees with financial assistance available for those in need. For more information, please visit www.medicalert.org or dial 888-633-4298 within the U.S. or 209-668-3333 from outside the U.S.

Alternatively, there are several companies that manufacture and inscribe medical identification jewellery. These products are available on the internet and are also carried by some jewellery stores and pharmacies. It may be wise to select jewellery which provides space for a miniature version of the patient's medical history (often supplied by the manufacturer as part of the cost) in addition to the inscription. Although these companies do not have a 24 hour information center, they do not require membership or annual fees and may be preferable for some patients.

It may also be helpful for mast cell patients to carry with them at all times emergency information, written on their physician's letterhead, which contains a list of current medications and other instructions for treatment in the event of severe symptoms.

DURING SURGERY

Constant attention from the anesthesiologist is required for the safety of a patient with mast cell disease during surgery, as some of the early symptoms of mast cell mediator release such as flushing, hives, and early signs of obstructed breathing can be masked by the surgical drapes covering much of the patient's skin and by the use of an airway tube during anesthesia.

Should anaphylaxis occur during surgery, the drug thought to be responsible should be discontinued immediately and epinephrine should be administered. Airway support with 100% oxygen, IV replacement fluids to compensate for dilated blood vessels, H1 and H2 antihistamines, bronchodilators, and corticosteroids may also be given. Continuous IV epinephrine and other "vasopressor drugs" may be necessary to keep blood pressure from falling. However, it is important to keep in mind that not all hypotensive episodes during surgery are due to mast cell degranulation and anaphylaxis. A serum tryptase level obtained during the hypotensive episode and its comparison

with pre-surgery or “baseline” level may be helpful to determine whether the episode is due to mast cell degranulation.

DRUGS

Records from prior surgeries should be examined and drugs tolerated in those procedures should be preferred if possible. Prior to the administration of any drugs associated with surgery, it is important that an IV is running, that epinephrine is available for immediate intramuscular (IM) or IV administration, and that emergency equipment is easily accessible in case of an adverse reaction.

IV preparations without preservative should be used.

Drugs to avoid include ethanol, dextran, and non-steroidal anti-inflammatory drugs (NSAIDs) such as ibuprofen and toradol (unless the patient is already successfully taking a drug in this family), polymyxin B, amphotericin B, quinine, dextromethorphan, α -adrenergic blockers, β -adrenergic blockers, and anticholinergic drugs as well as drugs mentioned in specific categories below.

Local anesthetics – True allergic reactions to local anesthetics resulting from mast cell degranulation are thought to be rare. Skin testing and graded challenge protocols have been published and may be administered by an allergist prior to the surgery if questions arise regarding the safe use of a local anesthetic in a patient. If possible, preservative-free local anesthetic should be used in these tests, as the preservative often present in local anesthetics can cause a mast cell reaction. In general, local anesthetics in the “ester” group should be avoided. This group includes procaine, chlorprocaine, tetracaine, and benzocaine. Anaphylactic reactions to local anesthetics in the “amide” group are rare. This group includes lidocaine, mepivacaine, prilocaine, bupivacaine, levobupivacaine, and ropivacaine.

Muscle relaxants - Muscle relaxants are the most likely group of the anesthetic drugs to cause anaphylaxis. Succinylcholine, D-tubocurarine, metocurine, doxacurium, atracurium, and mivacurium are more likely to cause a severe reaction than rocuronium or the so-called nondepolarizing muscle relaxants such as pancuronium or vecuronium. Some studies have reported increased numbers of anaphylactic reactions to rocuronium, however, so it may not be appropriate as the first choice for patients with mast cell disease.

Induction drugs - These are medications given to initiate anesthesia. It is rare for mast cell activation to occur in response to the use of propofol, ketamine, or the benzodiazepine drugs such as midazolam.

Inhaled anesthetics - Sevoflurane is an inhibitor of mast cell activation and is less likely to cause liver damage than other inhaled anesthetics in this family.

Opiates and opioids - Oral opioid drugs for pain relief may be tolerated by some mast cell disease patients, but their use should be approached with caution, beginning with very small doses. All drugs in this category are capable of causing mast cell mediator release.

SUMMARY

Surgery for a person with mast cell disease involves exposure to drugs and conditions that may trigger extensive mast cell degranulation. It should be emphasized that it is often impossible to predict or avoid the risk of adverse events which may occur in surgery due to the administration of drugs and the procedure itself. The risk can be reduced, though, if the mast cell disease is brought to the attention of everyone involved in the patient’s care and measures are taken to reduce the possibility of mast cell mediator release. Careful planning is important, beginning from the time a need for surgery has been defined and continuing through the immediate pre-operative period, anesthesia, surgery, and recovery. The choice of drugs is of major importance, and the anesthesiologist, the

surgeon, the nursing staff, the patient's physician, and the patient should form a working team to ensure that surgery presents the fewest possible dangers for the patient.

Neither The Mastocytosis Society nor the authors intend that this information replace medical advice given by the patient's doctor. Patients are encouraged to consult with their doctor regarding medications and procedures related to surgery.

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